CASE OF PROBABLE TUMOR OF THE PONS.[1]

By MARY PUTNAM JACOBI, M.D.

THE child whom I have the honor of bringing before the Society to-night has the following history.

Jennie Baer, aged ten years. The morbid symptoms are said to date from two and a half years ago, when the child was frightened by a trivial incident that she remembered with rather remarkable tenacity, and describes with lively interest. She was fastening the apron of a school friend, when the mother of the latter ran at her, uttering some injurious epithet. The child, the patient, ran away as fast as she could, up several flights of stairs to her own room, and immediately began to tremble. Two days later, on going to school, this trembling was noticed by the teacher. It continued, and even increased for two weeks, at the end of which time the teacher advised removal from school. She has never returned, nor attempted any systematic exertion since, bodily or mental. A little while after the removal from school, the parents noticed that the child walked as if intoxicated. The father imitates this mode of walking by slightly swaying the body from side to side, leaning a little forward, dragging one leg after another without lifting them from the floor, but somewhat swinging each in a semicircle, as if to enlarge the base of support. It is the walk "en fanchant" of the French writers.

At this time, and on account of the trembling and inability to use the hands for writing or other fine work, the parents consulted Dr. Lilienthal, who diagnosed St. Vitus' dance, and treated the child accordingly, but without benefit. At this time, though there was said to be twitchings of the limbs, there was none of the muscles of the face. The head was, however, habitually drawn a little to the left side, and the child exhibited the taciturnity common in chorea.

---

[1] Read before the Neurological Society, December 2, 1888.

After this period, *i. e.*, a month or two from the debut of the accidents, the child seemed constantly tired, and usually spent the time lying down. She played very little.

At the end of about three months the child began to have trouble with walking, would frequently stumble, and occasionally fall. Could not go up and down stairs readily. It was at the same time that she began to grow very fat.

She was brought to my clinic at the Woman's Medical College, in May of this year, 1888, after two years' continuance of the above symptoms. She had then been suffering during several weeks from headache, continuous, diffused, but by no means violent. It may be noted at this point, that this headache disappeared after a duration of seven or eight weeks, and has not returned. There had been two attacks of vomiting, at an interval of several weeks. These have not been repeated.

When the child was seen by me in May, I was at once struck by her large fleshy size, the unusual development of the limbs, and especially the great size of her head. The measurements taken at the clinic have been lost; but a month later, Dr. Petersen kindly made a careful examination of the head for me, and, together with some other determinations, the following measurements were taken:

Circumference equals 57⅓ centim., or 22⅗ inches.

Naso-occipital line equals 38 centimetres, or 15 inches.

Binauricular line equals 38 centimetres, or 15 inches.

I omit for the moment the other calculations made by Dr. Petersen.

I could not ascertain whether the mother had been impressed by the large size of the child's head, or whether she had noticed that it increased. The habitual expression of the child's face was apathetic; the eyes heavy and lids drooping; but if spoken to, her face lighted up and she replied intelligently and with a smile.

There was no hyperæsthesia of the scalp, and percussion of the head elicited no pain. There was no disturbance of sensibility of any kind in any part of the face, trunk or limbs. No sign of paralysis of any cranial nerve. The movements of the upper extremities were normal, though there was a little clumsiness in the finer movements of the fingers, as in picking up a pin. There seemed to be some paresis of the trunk muscles, at all events of the extensors of the spinal column; for the child avoided sitting upright, and continually leaned against the back of the chair for support. The conspicuous disturbance of function was in the lower extremities. The child could, though with some difficulty, rise from her chair and stand without support, but only for a minute or two, for she then fell forward. She could walk a few steps across the room, but then also would fall forward unless supported. During the attempt at walking, the body swayed a little from side to side, and the legs described the semicircular curves already mentioned. A month later it was noticed that the right ankle bent while walking, so that the foot turned inward. This was not perceived in May. At that time the child could walk up stairs more easily than on a level, because she supported herself by the bannisters. Faradaic sensibility and motility were intact for all muscles.

The knee-jerk was somewhat exaggerated on both sides.

The visceral functions were all normal; the appetite was even excessive. There was no unnatural drowsiness.

A re-examination was made of the case on October 13th.

The condition was found much aggravated in many respects. The circumference of the head has somewhat enlarged a little. Having been 57 centimetres, or $22\frac{3}{5}$ inches, when measured by Dr. Petersen in June, it is now $23\frac{1}{4}$ inches, a difference of $\frac{13}{20}$ of an inch. The naso-occipital and binauricular measurements remain the same.

The general appearance of the child is as already described; but she has grown still fatter, the girth of trunk, as measured by her dress which has burst out, being increased at least three inches.

The intelligence remains clear, notwithstanding the habitual listlessness. The child describes certain lines drawn on paper as vertical or horizontal, though she has not heard these terms since she left school two and a half years ago. She also relates the story of her original fright, and inquires with interest concerning the prognosis of her disease.

A new symptom now exists, in the intermittent divergent strabismus of the right eye. This is occasionally quite marked, but can always be overcome by voluntary effort, when converging the eyes to look at an object placed not too near. At a distance of two feet the nature and number of the objects are accurately distinguished; but when five lines are drawn on a paper, and the child looks closely at them, she calls them four. If she attempts to count the lines by placing her fingertip on them one after the other, the difficulty is increased. In six trials she invariably skipped the third line, and could not accurately touch the fifth. (They were drawn horizontally one above the other.) This difficulty depended partly on the inability to move the finger with precision; for it was agitated with slight choreiform movements during the intended act, which delayed its accomplishment, as is the case in sclerosis. But, in addition, there seemed to be some visual defect which interfered with the exact guidance of the finger and which rendered the patient unconscious of the fact that the finger had been placed above the line instead of on it. I could detect no diplopia; but I believe the lines seemed blurred to the child during the effort of convergence of vision upon very near and similar objects. This blurring she could not be made to describe.

There is no evidence of paralysis of the facial. The tongue protrudes perfectly straight. The uvula is slightly deviated to the right side. The tonsils are much enlarged.

Thus the only morbid symptoms in the sphere of the cranial nerves, apart from the optic, are: 1st, the intermittent irritation of the right abducens, as indicated by the intermittent divergent squint; 2d, the difficulty of converging the eyes upon near objects, so as to see them without blurring—difficulty apparently due to the relative or positive weakness of the right internal rectus.

The ophthalmoscopic examination was made later.

Upper Extremities.—The arms are very large; the forearms do not seem to be disproportionate to the age of the child. All movements of the arms and forearms can be executed without perceptible difficulty. Difficulty first appears when the child attempts to carry a glass of water to her mouth. Left to herself, she takes this in both hands, as if distrustful of her ability to hold it in one, although, on being tested, she can do so, and even, though unsteadily, carry it to her lips. When the glass is held by both hands against the mouth, she drinks readily until it is half emptied, but then seems unable to tip the glass at a greater angle in order to drain it. This movement necessitates the inclination of the hands on the radial border of the forearms, action effected by the supinator longus, and the longer and shorter radial extensors of the wrist. When there is no weight in the hand, the child can flex and deflect the hand on the radial border of the wrist; the difficulty only occurs when the hand is carrying a weight and for a special purpose. There is therefore no actual paralysis of the muscles, but paresis and diminished power of co-ordination.

A similar difficulty of co-ordination is show in the fingers when the child tries to write. Before the hand reaches the pen it is agitated with slight choreiform or ataxic movements, as is also the case when she picks up a pin. She cannot go straight to the object. She places her head to one side, the right,in order to guide her hand in writing. She then forms the letters very imperfectly, and cannot write them in a straight line. The word (she resigns the attempt after the first word) always runs obliquely off the page, running up sharply from left to right. It is to be remembered, in connection with this test, that the child's education ceased when she was only eight years old, and before she had really learned how to write. Apart from efforts at functional co-ordination, all movements of the fingers can be performed voluntarily and without distinctly perceptible diminution of force. The grasp of the hand is normal for a child of her age. Sensibility is intact for the whole upper extremity.

The faradaic contractility and sensibility are intact.

Trunk.—The child cannot support herself upright, even in a sitting position, for more than a minute or two, but leans back in the chair, showing paresis of the spinal extensor muscles.

The thighs and legs are noticeable for their large size, although the apparent muscular enlargement of the four extremities is not as striking as is that of the thorax and abdomen. While sitting, the child can extend the legs on the thighs perfectly, but not vigorously, and can flex, extend, and adduct the feet and toes. Abduction is extremely feeble on both sides. Nevertheless, she is unable to support herself standing for more than a half minute, and cannot now walk at all unless supported. With support she walks, dragging and to some extent swinging round the legs, while both ankles bend under her, bringing the feet into exaggerated varus. Thus there has been marked deterioration in the functional power of the limbs since May, and even since June, when the special weakness of the peroneal was first observed, and then only on the right side.

There is no retraction of the gastrocnemii muscles. The child has never walked on the toes, nor never exhibited the special phenomenon of pushing back the thighs or of climbing up them in rising. Nor was there ever lordosis, but the trunk always fell forward.

In May, as already stated, the child did pull herself up stairs by the aid of her arms. Also, while she was yet able to walk unsupported, the body did sway from one side to the other, as if from alternate inclination of the pelvis, and the legs spread outwards to enlarge the base of support. These symptoms have now disappeared, as the paresis has increased so far that the child cannot even attempt to walk unsupported.

The absence of retraction in the gastrocnemii is parallelled by similar conditions throughout the limbs, where neither contracture nor deformity exists. The sensibility is intact. All the muscles contract to the faradaic current, but the peroneal muscles demand a stronger current than the others. Thus the quadriceps extensors contract at 300 millimetres of the induction coil, the peroneal not under 260. The knee-jerk is moderately exaggerated. No ankle clonus can be obtained.

The limbs are constantly cold, subjectively and to palpation.

In addition to the foregoing symptoms may be noted:

That the child suffers no pain in any part of the body, nor has ever done so except for a moderate headache in last spring.

There is neither nausea, vomiting, nor anorexia, but, on the contrary, an excessive appetite, bordering on bulimia. There is occasional sighing respiration.

There is no constipation, but a condition of the bowels approaching incontinence, inasmuch as the child is unable to restrain the impulse for evacuation when this makes itself felt; and the impulse always occurs after any mental excitement, as if she has been laughing. The control of the bladder is similarly imperfect.

On the 28th of October Dr. C. S. Bull kindly made an ophthalmoscopic examination of the child. He found choked disk in both eyes, outlines much blurred, veins swollen and tortuous, arteries narrowed and in some places disappeared; process beginning to recede,but leaving an atrophy of nerves, which is now incipient, but tending to increase Acuity of vision much diminished, only $\frac{10}{100}$. Spasm of right abducens. He diagnosed a descending inflammatory process of the optic nerve.

In 1887 Dr. Hun, of Albany, presented the following case, completed by the autopsy, to the American Neurological Association:

Gliomatous hypertrophy of the pons.

Female, æt. 6; father died a little more than a year after she was born, with symptoms of melancholia and dementia.

For several years left leg has tired easily; left foot shows a tendency to turn in; wore rubber straps; otherwise excellent health until two months ago, when attack of croup and cough. Pain felt in head with each cough. Three weeks before coming under observation the patient began to walk badly, and seemed to have trouble in balancing herself while walking. She had an excessive appetite and vomited a little at times.

When first seen the patient was a well-nourished, intelligent girl, but with a vacant expression. Her speech was drawling. Her head was drawn towards the right shoulder most of the time, especially if she made any exertion. She stood with her feet wide apart, and was careful not to lose her balance. In walking, her right leg was more rigid than her left, so that she took freer and longer steps with her left leg, and therefore in walking tended to go in a circle, turning always to the right. Her walk resembled that of a drunken person. The movements of the arms, especially of the right, were very awkward, but she held them in no fixed position.

There was no disturbance of sensibility in any part of the face, body, or extremities, and she recognized objects placed in her hands when her eyes were shut.

The plantar reflexes were normal. The knee-jerk was exaggerated, especially on the right side. There was no ankle clonus.

On ophthalmoscopic examination, well marked optic neuritis was found in both eyes. Urine contained neither albumen nor sugar.

The patient was first seen on the 17th of April, and from this time the symptoms rapidly increase, passing through gradations I need not here describe, until a fatal termination was reached on the 13th of June.

At the autopsy, the pons varolii was found enlarged to three or four times its normal size, and on section, it was found to have been replaced by a tumor, apparently a glioma, which so well preserved the normal appearance of the part that it looked like a greatly hypertrophied pons.

On microscopical examination, the nervous elements of the pons were found to be encroached upon, infiltrated, and in some places destroyed by a great accumulation of small cells. The proliferation of cells was not limited to the pons, but extended throughout the crura cerebri and medulla, and especially in the roof of the aqueductus Sylvii. The prevailing character of the cells was spindle, but there was a considerable number of spheroidal cells, both large and small.

The bones of the skull were thin. The subarachnoid fluid increased. The cerebral convolutions flattened, and the cerebral substance very œdematous. Lateral ventricles greatly dilated, and filled with fluid of normal appearance.

Dr. Hun ascribes the inco-ordination of movements to pressure upon the transverse fibres of the pons, and the origin of the crura cerebelli; the absence of absolute paralysis to the fact that the nerve elements were compressed but not destroyed by the infiltration. He explains the absence of sensory disturbance by a greater resistance of sensory function, even when sensory fibres are submitted to the same pressure as motor fibres. He notes that the tumor in its growth produced no symptoms of irritation, that there were no convulsions, and but little headache.

In all these circumstances, the case closely resembles the one I now present. Other points of resemblance are the long duration of slight muscular inco-ordination, and even paresis of the lower extremities, without other symptoms, up to a certain date; then the rapid deterioration in a few months; the fact that the most marked paresis was shown by the turning in of the right ankle; the exaggeration of the knee-jerk, the other reflexes remaining normal; the vacant expression of face, and apathetic appearance, although the memory and intelligence were entirely preserved; the indistinctness of articulation; the optic neuritis, the slight and transitory squint, which in Dr. Hun's case was convergent, in this one divergent and unilateral, without paralysis of any ocular muscle; finally, the excessive appetite and excellent preservation of nutrition. The nutrition in our case is even exaggerated, so as at one time to have suggested a general lipomatosis.

The optic neuritis is, as well known, a most important indication of intra cerebral tumor; and indeed I did not permit myself to make a positive diagnosis until the existence of this symptom had been established. Bramwell enumerates the following diseases other than mental in

---

[1] A case of tumor of the pons and medulla in a child of two years, related by Hobson, (Brain, Vol. IV., p. 531), differed greatly from ours, especially in the number of cerebral nerves involved.

which double optic neuritis may occur. It is nearly always present in lead encephalopathy; it is not uncommon in meningitis and cerebral abscess; and it may occur in Bright's disease. It has occasionally been associated with uterine derangements, hypermetropia and anæmia, which, when complicated with hysterical symptoms, may be easily mistaken for cases of intra cranial tumor.

In our case all the foregoing conditions may be readily excluded.

Stephen Mackenzie has published in the second volume of Brain, a case where an optic neuritis depended upon a diffused cerebritis, resulting in general atrophy of the brain, and an inflammation extending down the optic nerve. The symptoms of this remarkable case, which has often been quoted, are chiefly to be referred to the cortex of the brain, and in no wise resembled those of this child.

When I first saw the patient, the unusually large size of the head, the apathetic expression of face, the moderate degree of motor disturbance, the diffused nature of the symptoms, led me to suspect an hypertrophy of the brain, due to diffuse lobar sclerosis. "In such cases," observes Gowers, "the symptoms have been very similar to those of cerebral tumor, headache, vomiting, local palsy, convulsions."[3]

"The symptoms," observes Schmidt,[4] "of diffused sclerosis of the brain are gradually increasing muscular weakness, manifesting itself especially in the lower extremities, giving rise to an unsteady, stumbling gait and frequent falling. There are also epileptic spasms, constant or intermittent attacks of headache, vertigo, tinnitus aurium, photophobia, dimness of sight with dilatation of the pupil, blunting general sensation without anæsthesia. The skull may be enlarged and thinned."

So Richardière[5] affirms: "Convulsions and attacks of tremor and muscular rigidity are never absent in cases of diffused sclerosis." Both these symptoms have been entirely lacking in the case of Jennie Baer. On this account the hypothesis of the diffuse sclerosis was finally rejected.

---

[3] Diseases nervous system.

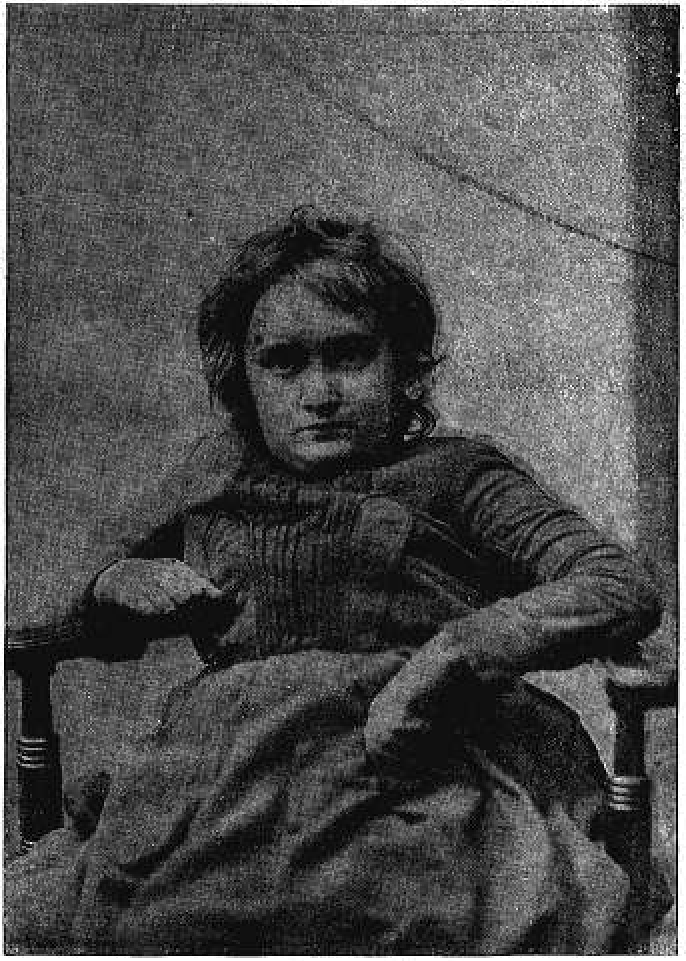
[4] Peppers Archives of Medicine.

[5] Scleroses encephaliques primitives de l'enfance. Paris, 1885.

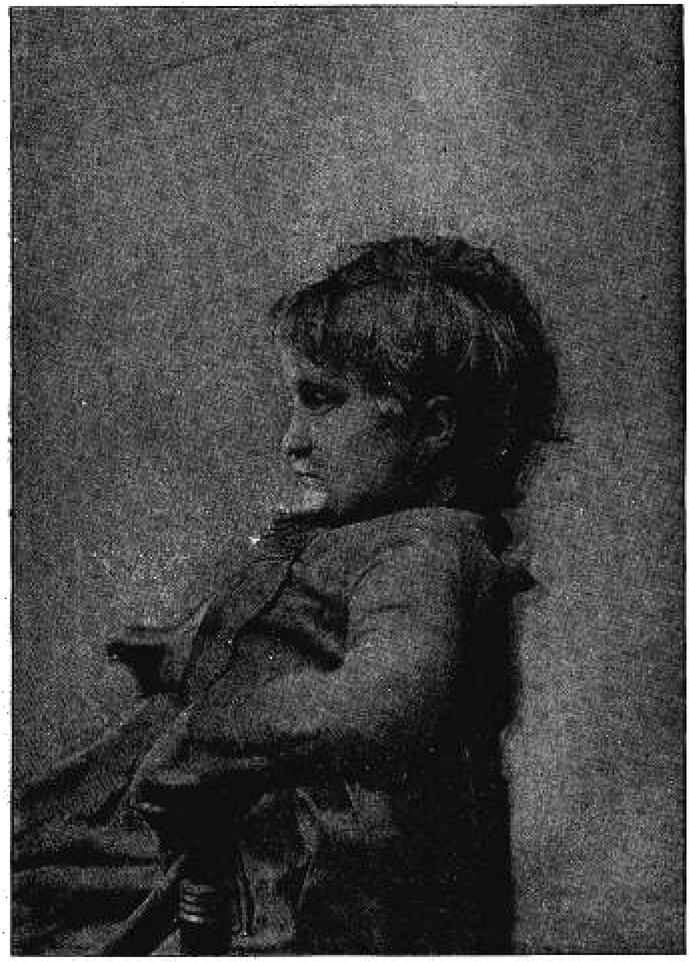
To sum up: the reasons for diagnosing an intracranial tumor in the case of the child before you, are: 1st. A diffused motor disturbance, beginning as inco-ordination and difficulty of equilibrium, increasing at first slowly, then with sudden rapidity to such paresis of the lower extremities as renders station impossible; of the trunk muscles, as renders upright sitting difficult; of the upper extremities, as interferes with the more delicate movements of the hands. 2d. Preservation of faradaic contractility, absence of nutritive lesion of the paretic muscles. 3d. Gradual enlargement of the head. 4th. Moderate apathy of expression and dulling of intelligence. 5th. Spasm of right external rectus. 6th. Double optic neuritis, commencing atrophy of the optic nerves, marked diminution of visual acuity.

The circumstances which indicate a localization of the tumor in the pons are: 1st. The absence of convulsions, a negative fact of great importance, and frequently observed in slowly infiltrating tumors of the pons. 2d. Absence of marked or definite symptoms, and of monoplegic spasm or paralysis, tend to exclude tumors of the cortex. 3d. Absence of hemiplegia excludes the basal ganglia. 4th. The same, with absence of symptoms in the sphere of the motor oculi, excludes tumors of the crura or base of the brain. 5th. Absence of headache, nystagmus and vomiting, and the development of motor paralysis in addition to the original motor inco-ordination, excludes, I think, the cerebellum. 6th. The general march of the symptoms, the bilateral character of the paresis, the inco-ordination, even the absence of anæsthesia or pain, are precisely what have been observed in slow growing tumors of the pons. In seven out of thirty cases of pontine tumor tabulated by Bernhardt, no disturbance of sensibility existed but headache, and even this failed in two cases, as also in that of Dr. Hun. The irritation of the right abducens, the only cranial nerve at present affected except the optic, is in accordance with a localization of disease in the pons. The excessive appetite of the child is a symptom probably to be referred to the medulla.

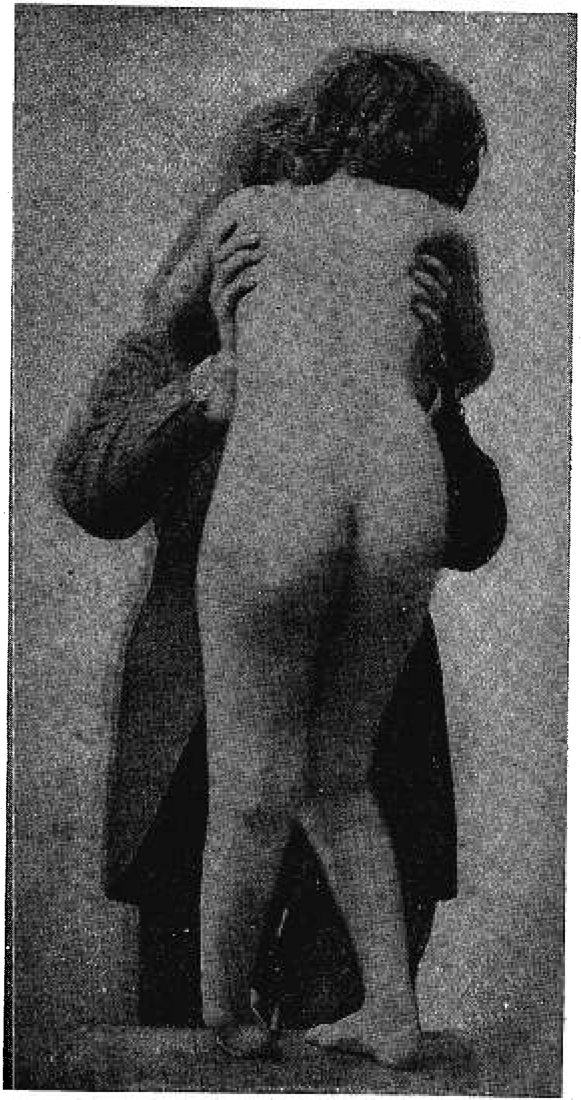
Characteristic symptoms of pontine tumors which are, as yet, absent, are: 1st. Alternate paralysis of face and limbs. 2d. Paralysis of the hypoglossus, or of any cranial nerve, other than the optic. 3d. Marked difficulty of deglution or articulation. Some defect of the latter function is, however, noticeable, and there is occasionally sighing respiration. The tumor could hardly be situated in the upper



region of the pons, or there would be motor oculi symptoms, or others referring to the corpora quadrigemina; while at the lowest portion of the pons, the spinal accessory nucleus or the hypoglossal nerve should be involved. We may



infer that the growth is situated about the middle region, that it is bilateral, that it is below and anterior to the nucleus of the fifth and anterior to the nucleus of the seventh nerve. That on this account the lower extremities are paralyzed before the upper, and the seventh has so far escaped. We must further infer that the lesion consists of



a very gradual infiltration of elements, gliomatous or sarcomatous into the nerve tissue, and is not a sharply defined new growth, forciby compressing any localized bundles of nerve fibres.

One other hypothesis remains,[6] namely, that there is a tumor of the cerebellum which presses on the pons. The reasons which militate against this hypothesis are, that there have never existed any characteristic symptoms of cerebellar tumor, except the tottering walk and loss of equilibration. But these symptoms are also observed in the infiltrations of the pons which affect the transverse crura cerebelli.

In the four cases of cerebellar disease reported by Dr. Seguin,[7] the following symptoms, positive and negative, were observed, differing from the history of Jenny Baer.

CASE I.—*Positive:* Headache, nystagmus, convulsion. *Negative:* Absence paralysis until after hæmorrhagic seizure two months before death.

CASE II.—*Positive:* Vomiting, violent headache, convusions.

CASE III.—Diffused headache, vomiting, double exophthalmus, absence paralysis.

CASE IV.—Repeated nausea and vomiting, severe occipital headache, absence paralysis.

The paralysis in the case of Jenny Baer is not indeed complete, but it is sufficient in the lower extremities to render standing quite impossible, which is more extensive than is ever the case with purely cerebellar tumors.

---

[6] This was strongly urged by Dr. Sachs at the meeting of the Neurological Society when the above communication was read.

[7] Journal Nervous and Mental Disease, vol. xiv., April, 1887.